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SHORT REPORT

Inferior Vena Cava Agenesis with Paravertebral Muscle Collateralisation

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Introduction

Developmental anomalies of the inferior vena cava (IVC) have an estimated prevalence of 0.5%, and may be the common aetiology in young patients presenting with bilateral lower limb iliofemoral thrombosis.[1–3] We report a case of spontaneous bilateral iliofemoral deep venous thrombosis secondary to infrahepatic IVC agenesis. The patient had associated thrombosis of the azygous and hemiazygous venous system and collateralisation via the venous plexus of the paravertebral muscles. To our knowledge, this is the first such case reported of IVC agenesis dependent upon collateral return via the paravertebral muscles.

Case Report

A 21-year-old male was admitted with a 10 day history of increasing pain and bilateral lower limb swelling. The patient had no known risk factors for venous thrombosis, in particular he was a non-smoker, with no recent flight history and no personal or family history of thrombosis. The lower limbs were found to be grossly swollen bilaterally from both ankles to the thighs and were associated with calf tenderness. All pedal pulses were palpable. There was no clinical evidence of pulmonary embolism.

A clinical diagnosis of bilateral deep venous thrombosis was made. The patient was anti-coagulated with intravenous Heparin. Doppler ultrasound (colour and compression B mode) demonstrated extensive occlusive thrombus involving the distal external iliac veins and the common and superficial femoral veins bilaterally extending to the popliteal fossa. A computerised tomogram (CT) (iv contrast, spiral scan of abdomen), revealed thrombosis of the common femoral and iliac veins and gross dilatation of the upper parts of the azygous and hemiazygous veins. This was consistent with an IVC obstruction and venous return in the lower abdomen occurring mainly via deep muscular branches (Fig. 1). There was also absence of the infra hepatic IVC (Fig. 2). A magnetic resonance image (MRI) demonstrated a collateral deep venous plexus in the para vertebral muscles (Fig. 3). The patient was anticoagulated with Enoxaparin (Clexane, Aventis) 100 mg b.d., and subsequently commenced on Warfarin. A thrombophilia screen revealed no other predisposition to thrombosis. He had no further thrombosis at 1 year follow-up.

Discussion

The overall incidence of bilateral deep venous thrombosis in the population is 0.08–8.6%.[4,5] However, in the presence of IVC anomalies the incidence of bilateral thrombosis may be as high as 62.5%.[3] The IVC develops during embryogenesis via development, regression and anastomosis of three sets of paired veins: the posterior cardinal, sub-cardinal and supra-cardinal veins.[6] The normal inferior vena cava converts to a unilateral right-sided system consisting

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Fig. 1. Venous return via deep muscle venous plexus.

of four segments the hepatic, pre-renal, renal and post-renal segments. Failure of these structures to unite results in anomalies of the IVC in up to 0.5% of the general population.[1]

Ruggeri and Chee, using CT and MRI respectively, have identified IVC anomalies in patients presenting with extensive deep venous thrombosis (DVT).[2,3] Both authors noted that such thrombosis occurred in a younger age group than expected normally for venous thrombosis. Obernosterer *et al*. carried out a prospective analysis of 97 patients with DVT. Five patients demonstrated anomalies of the inferior vena cava on magnetic resonance angiography.[7] These studies indicate the importance of considering such IVC anomalies as independent risk factors for DVT formation. This unusual aetiology should be particularly considered in young patients, with no other risk

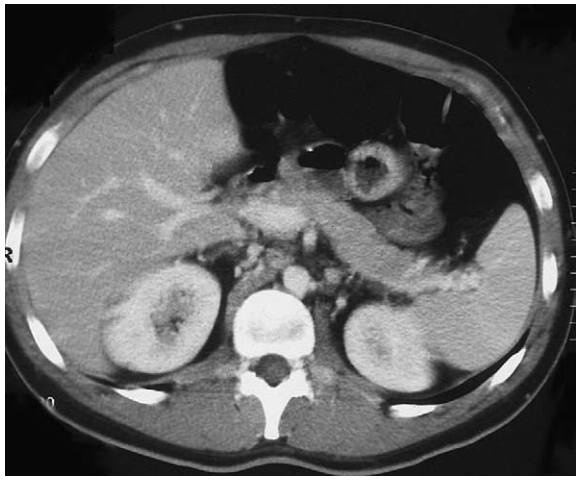

Fig. 2. CT scan showing aorta and absence of normal infrahepartic inferior vena cava.

Fig. 3. T1 weighted MRI showing aorta and extensive retroperitoneal venous channels.

factors, presenting with bilateral disease. This case highlights the importance of including abdominal scanning when dealing with this unusual subgroup of patients.

Despite the massive nature of his thrombus, we believe our patient to be at low risk of developing a pulmonary embolism. Paradoxically, his dependence on the small deep veins in the para vertebral muscles for venous return may have conferred anatomical protection from pulmonary embolism. Currently, no guidelines exist as to the correct management of such patients. Life long anticoagulation appears prudent, however, the long-term necessity and the duration of anticoagulation in such a young age group are unclear. In our patient, long-term anticoagulation was felt necessary due to the severity of his presentation and to possibly prevent chronic venous hypertension. However, this may be considered by some to be over cautious.

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